

# Challenging The 2015 PH Guidelines - comments from the Nurses



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# Recommendations for pulmonary hypertension expert referral centres

Recommendations	Class <sup>a</sup>	Level <sup>b</sup>
It is recommended for expert referral centres to provide care by a multiprofessional team (cardiology and respiratory medicine physicians, clinical nurse specialist, radiologists, psychological and social work support, appropriate on-call expertise)	I	C
It is recommended for expert referral centres to have direct links and quick referral patterns to other services (such as CTD, family planning, PEA, lung transplantation, adult congenital heart disease)		
It should be considered that a expert referral centre follow at least 50 patients with PAH or CTEPH and should receive at least two new referrals per month with documented PAH or CTEPH		
It should be considered that a expert referral centre perform at least 20 vasoreactivity tests in IPAH, HPAH or DPAH patients per year	IIa	C
Expert referral centres should participate in collaborative clinical research in PAH, including phase II and phase III clinical trials	IIa	C

## Pulmonary arterial hypertension therapy

It is recommended for expert referral centres to provide care by a multi-professional team (cardiology and respiratory medicine physicians, clinical nurse specialist, radiologists, psychological and social work support, appropriate on-call expertise)

I

C

# 12.1 Facilities and skills required for an expert referral centre

Expert referral centres are recommended to provide care by an interprofessional team that should, as a minimum, comprise:

- (a) two consultant physicians (normally from either or both cardiology and respiratory medicine) experienced in and with a special interest in PH with dedicated PH clinical sessions for outpatients, inpatients and a multidisciplinary team meeting
- (b) clinical nurse specialist
- (c) radiologist with expertise in pulmonary hypertension imaging
- (d) cardiologist or PH physician with expertise in echocardiography
- (e) cardiologist or PH physician with expertise in RHC and vasoreactivity testing
- (f) access to psychological and social work support
- (g) appropriate on-call cover and expertise

# Clinical Nurse Specialists

5 main competency areas<sup>1</sup>:

- Clinical expertise
- Research
- Leadership
- Collaboration
- Change agent

Demonstrated by<sup>1</sup>:

- Listening , counselling
- Supporting patients and families
- Providing direct care
- Formal education
- Collaboration with the healthcare team
- Advocating for additional resources for patients and family members

# The impact of CNSs on patients living with a chronic disease

- Improved quality of life <sup>1 2 3 4</sup>
- Fewer and shorter hospitalisations <sup>1 5 6 7</sup>
- Decreased costs of care <sup>1 7 8 9</sup>
- Increased patient and health provider satisfaction <sup>1 8 9 10</sup>

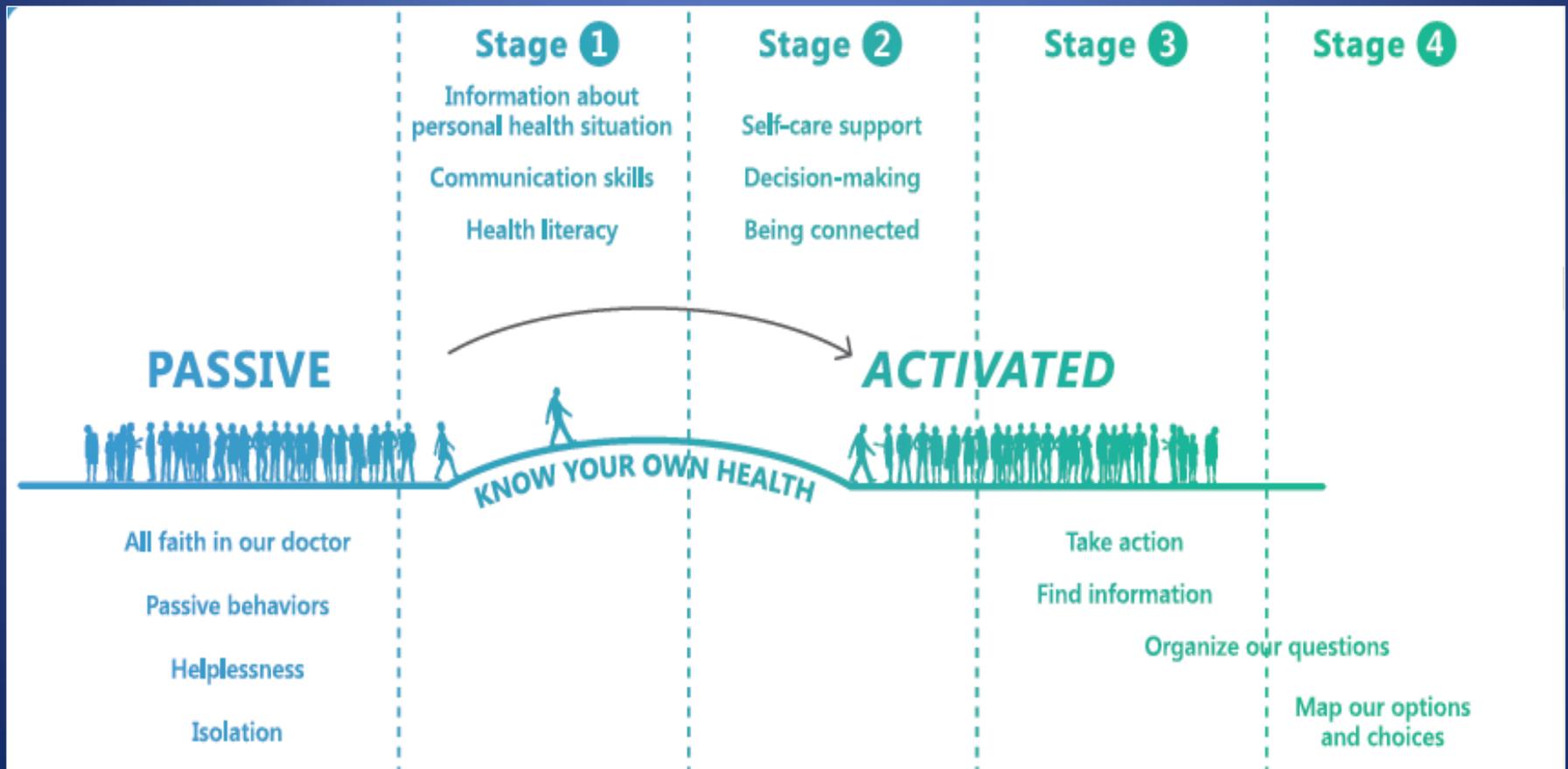
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2. Ritz (2000) Oncol Nurse Forum. 27 p9233
3. Kutzleb (2006) J Am Acad Nurse Pract. 18 p116
4. Maughan (2001) J Clin Nurs. 10 p221
5. Naylor (1999) Eur J Cardiovasc Nurs. 14 p44

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# Advances in the management of pulmonary hypertension

- Over the past 20 years PAH has transitioned from being a fatal disease with a poor prognosis to a long-term condition that, with the right treatment and support, can be managed.<sup>1</sup>
- Patient care delivered via specialist, multidisciplinary healthcare teams now has a greater focus on quality of life considerations alongside the clinical symptoms of the disease

# Patient engagement and self-management should be key considerations in PAH care



# Patients that feel well informed cope better with their disease



PAH patients who were **satisfied with the received information** found it easier to cope with their disease ( $p = 0.0045$ )



Those who **found the overall information useful** found it easier to cope with their disease ( $p = 0.003$ )

# Patient perception of received information

Results from a questionnaire for PAH patients ( $n = 68$ ) in Sweden

Information area	% of patients receiving info (n=68)
Disease	58
Medical tests	67
Treatments	47
Other services	21
Different places of care	23
Ways to help yourself	33

- **54% of PAH patients do not feel adequately informed about their disease**

# Areas in which patients desired more information

Topics in questionnaire	Other topics raised by patients
Diagnosis	Survival estimates/prognosis
Whether disease is under control	Research / future treatments
The severity of the disease	Possibility to become cured
Things patients can do to get well	Future palliative care
Side effects of treatments	Own hospital records
Purpose and results of medical tests	Co-morbidity
Possible causes	Risks of surgery
Rehabilitation services	Appropriate form of exercise
Psychological support	Unstressed, honest, straight forward information without having to ask and easy to get
Individually written information	

Based on questionnaire for PAH patients ( $n = 68$ ) in Sweden

# The optimal patient journey: Effective communication throughout the care process

	Patient information
First clinic visit	<ul style="list-style-type: none"><li>• Mechanisms of disease</li><li>• Investigations required</li><li>• Contact details of the PH Centre</li></ul>
Diagnosis	<ul style="list-style-type: none"><li>• On disease, treatment options, specialists involved in PAH care</li><li>• Managing the emotional impact; support via patient associations</li><li>• Written materials for patient to take away</li></ul>
Appointment 1 after diagnosis	<ul style="list-style-type: none"><li>• Written care plan following shared decision-making</li><li>• Patient 'passport' with individualised content</li><li>• Further information from patient association</li></ul>
Appointment 2 after diagnosis	<ul style="list-style-type: none"><li>• Referral to specialists as needed e.g. psychiatry, welfare</li><li>• Patient-to-patient mentoring matched according to age/background etc.</li></ul>
Appointment 3 after diagnosis	<ul style="list-style-type: none"><li>• Self-management course to support rehabilitation</li><li>• Ongoing engagement with multidisciplinary healthcare team and patient associations</li></ul>

# ESC Guidelines treatment goals

The overall treatment goal in patients with PAH is achieving a low risk status

**Table 13** Risk assessment in pulmonary arterial hypertension

Determinants of prognosis* (estimated 1-year mortality)	Low risk <5%	Intermediate risk 5–10%	High risk >10%
Clinical signs of right heart failure	Absent	Absent	Present
Progression of symptoms	No	Slow	Rapid
Syncope	No	Occasional syncope <sup>b</sup>	Repeated syncope <sup>c</sup>
WHO functional class	I, II	III	IV
6MWD	>440 m	165–440 m	<165 m
Cardiopulmonary exercise testing	Peak VO <sub>2</sub> >15 ml/min/kg (>65% pred.) VE/VCO <sub>2</sub> slope <36	Peak VO <sub>2</sub> 11–15 ml/min/kg (35–65% pred.) VE/VCO <sub>2</sub> slope 36–44.9	Peak VO <sub>2</sub> <11 ml/min/kg (<35% pred.) VE/VCO <sub>2</sub> slope ≥45
NT-proBNP plasma levels	BNP <50 ng/l NT-proBNP <300 ng/l	BNP 50–300 ng/l NT-proBNP 300–1400 ng/l	BNP >300 ng/l NT-proBNP >1400 ng/l
Imaging (echocardiography, CMR imaging)	RA area <18 cm <sup>2</sup> No pericardial effusion	RA area 18–26 cm <sup>2</sup> No or minimal, pericardial effusion	RA area >26 cm <sup>2</sup> Pericardial effusion
Haemodynamics	RAP <8 mmHg CI ≥2.5 l/min/m <sup>2</sup> SvO <sub>2</sub> >65%	RAP 8–14 mmHg CI 2.0–2.4 l/min/m <sup>2</sup> SvO <sub>2</sub> 60–65%	RAP >14 mmHg CI <2.0 l/min/m <sup>2</sup> SvO <sub>2</sub> <60%

6MWD = 6-minute walking distance; BNP = brain natriuretic peptide; CI = cardiac index; CMR = cardiac magnetic resonance; NT-proBNP = N-terminal pro-brain natriuretic peptide; pred. = predicted; RA = right atrium; RAP = right atrial pressure; SvO<sub>2</sub> = mixed venous oxygen saturation; VE/VCO<sub>2</sub> = ventilatory equivalents for carbon dioxide; VO<sub>2</sub> = oxygen consumption; WHO = World Health Organization.

# Treatment Goals

- Clear parameters - easy to assess
- BUT...
- Many patients remain in the intermediate zone despite increases in PH therapy
- There are many other factors which can influence parameters
- Many goals are not meaningful for patients

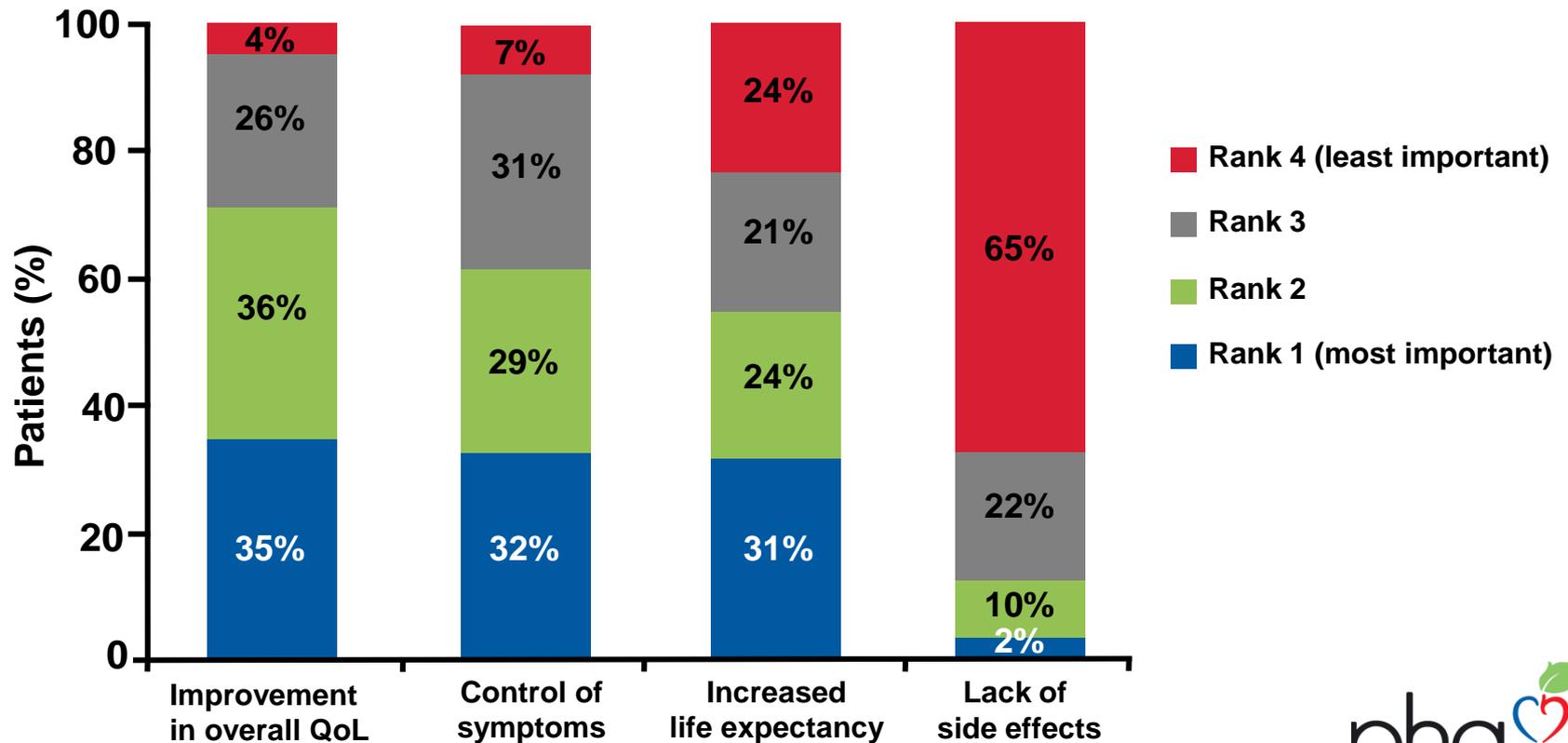
# What do Patients Want?

- To get an accurate diagnosis and the best treatment without avoidable delay
- To be treated with humanity, dignity and respect
- Information and communication
- Psychological, social and spiritual support
- Optimal symptom control
- Rehabilitation
- Well coordinated care
- End of life care
- Support for carers and bereavement



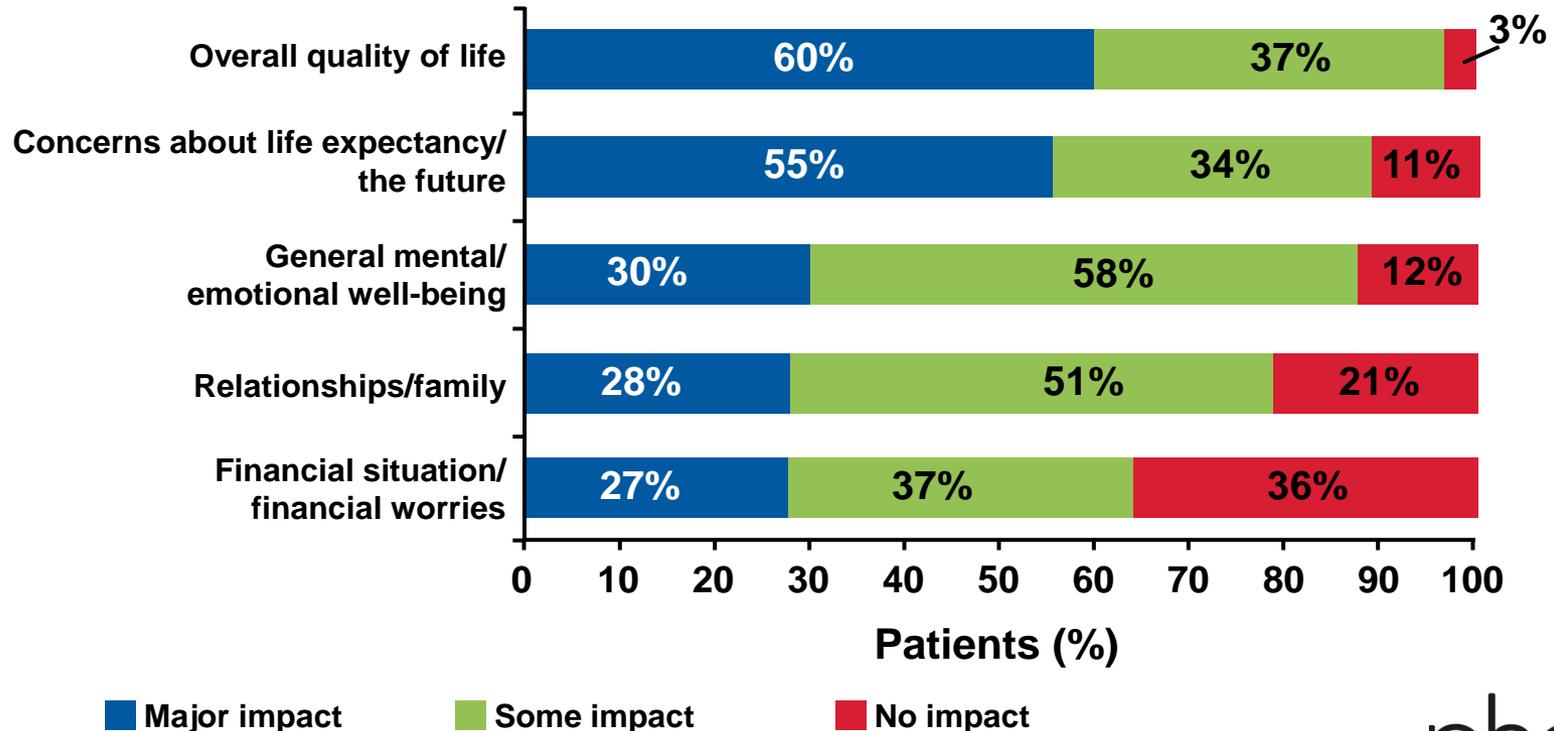
# Important factors when choosing treatments

- Majority of patients think QoL is the most important factor when choosing treatment



# Impact of PAH on a patient's life

- Overall QoL and concerns about life expectancy and the future have the biggest impact on the life of a patient with PAH



# Individualised goal setting

- Coaching patients to identify their own realistic, meaningful and achievable goals
- Develop a plan to achieve their goals
- Ongoing assessment, re-evaluation and setting of new goals

I want to be able to work and support my family

I want to live to see my granddaughter born

I want to go swimming with my children

I want to be pain free

## 6.3.1.5 Psychosocial support

PH is a disease with a significant impact on the psychological, social (including financial) emotional and spiritual functioning of patients and their families.

Teams managing these patients should have the skills and expertise to assess and manage issues in all of these domains, with close links to colleagues in relevant disciplines for those with severe problems, e.g. psychiatry, clinical psychology, welfare and social work.

# Depression

Because of diminished quality of life, decreased exercise capacity and mobility, social isolation, financial burdens, unemployment, and an uncertain prognosis, patients with PAH may be more vulnerable to developing depression<sup>1</sup>

Resulting in:

- Medical non-compliance<sup>1</sup>
- Increased levels of IL6<sup>1 2</sup>, CRP<sup>1 2</sup>, endothelial dysfunction<sup>3</sup> and BNP<sup>1</sup>
- Decreased 6 minute walk test distance<sup>1</sup>
- Decreased life expectancy<sup>4</sup>

1. McCollister (2010) Psychosomatics. 51 p339

2. Miller (2002) Am J Cardiol. 90 p1279

3. Rumsfeld (2003). J Am Coll Cardiol. 42 p1811

4. Ross (2012) BMJ. 345

## 6.3.11 End of life care and ethical issues

The clinical course of PH is one of progressive deterioration interspersed with episodes of acute decompensation. It is difficult to predict when patients will die since death may come either suddenly or slowly because of progressive heart failure. It has been shown that physicians caring for patients tend to be overoptimistic in their prognostication and frequently misunderstand their patients' wishes.

Open and sensitive communication with patients allows advanced planning and discussion of their fears, concerns and wishes, which is essential to good care.

# Palliative care in PH

- Many patients have a significant symptom burden despite optimal therapy
- Can be used in parallel with PH therapy to improve symptoms and provide support for patients and their carers
- Should be introduced earlier in the course of the disease as a component of standard care
- Has been shown to improve depression and increase survival in other diseases <sup>1</sup>

# Conclusion

- Patients should be encouraged to accumulate knowledge and skills to become empowered to live well with PAH
- Successful chronic disease interventions usually involve a coordinated multidisciplinary care team <sup>1</sup>
- CNSs can provide high quality and cost effective care to patients and families through education, support, advocacy and coordination of care <sup>2</sup>

1. Wagner (2000) BMJ.320 p569

2. Moore (2012) Clinical Nurse Specialist. 26(3) p149

# Thank you

